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Case Report

Postsplenectomy sepsis: a lifelong risk

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Overwhelming postsplenectomy infection is a term used to describe the clinical syndrome of fulminant bacteraemia, disseminated intravascular coagulation, multi-organ failure, severe hypoglycaemia and rapid death which has been described in patients who have been splenectomised for a variety of reasons.[1,2] Whilst the highest mortality has been reported in the early post-operative period and in the immunosuppressed[3], the following cases illustrate the fact that splenectomy carried out at any age and for any reason increases the risk of overwhelming bacterial infection. Therefore vigilance and continuing education are always required on the part of patients and their physicians.

CASE 1

A 63 year old male had a splenectomy performed in childhood following a road traffic accident. He felt slightly unwell, developed a temperature and became confused and agitated. Some 12 hours after the onset of confusion he was admitted to hospital. He was not taking prophylactic antibiotics and there was no record of pneumococcal vaccination. He was allergic to penicillin.

On admission his temperature was 38°C. He was noted to be very confused and agitated. There were no localising signs. He was commenced on erythromycin, and cefotaxime was then added.

Coagulation screening tests on admission showed disseminated intravascular coagulation which progressed despite attempts at correction. Deteriorating blood gas measurements necessitated intubation and ventilation, but despite continued aggressive resuscitation he died within 24 hours. Blood cultures taken before commencement of antibiotics identified streptococcus pneumoniae.

CASE 2

A 37 year old male had had a splenectomy when aged 13 years following abdominal trauma. He presented to hospital with a three day history of feeling feverish and vaguely unwell. He was not taking antibiotics and there was no record of vaccination. On examination his temperature was 39°C, blood pressure

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was 105/65mmHg and there was a widespread purpuric rash. Coagulation screening tests showed disseminated intravascular coagulation. Therapy was instituted with benzylpenicillin, dobutamine and fresh frozen plasma. Pre-antibiotic blood cultures identified *streptococcus pneumoniae.* He gradually improved but developed gangrene of the feet and all fingers, necessitating bilateral below-knee amputations and amputation of all fingertips. On pathological examination of the amputated limbs occlusion of the small vessels was confirmed consistent with a diagnosis of disseminated intravascular coagulation.

CASE 3

A 62 year old male had had a splenectomy in 1977 at age 47 for an acute haemolytic crisis in association with hereditary spherocytosis. He was found at home confused and faecally incontinent. Relatives gave a history that he had been vaguely unwell for several days. There was no record of vaccination and the patient was not taking prophylactic penicillin. On admission to hospital he was confused and obviously unwell, temperature 38°C, blood pressure 80/50mmHg. There were no localising signs. Despite intensive resuscitation including intravenous penicillin, gentamicin, metronidazole and ciprofloxacin he deteriorated. Disseminated intravascular coagulation present on admission improved with intervention, but he remained hypotensive and developed anuria. Increasing hypoxia and acidosis necessitated intubation and ventilation but he continued to deteriorate and died within 15 hours of presentation. No organism was identified from pre-antibiotic blood cultures or from post-mortem bacteriological examination. Autopsy confirmed the presence of a consumptive coagulopathy with bilateral massive adrenal haemorrhage and it was concluded that septicaemia was the cause of death.

DISCUSSION

The incidence of overwhelming postsplenectomy infection varies according to the study but has been reported as high as 6.9%,[4]. Although more common in the three years following surgery, sepsis can occur at any time. In case 1 more than 45 years had elapsed from the time of splenectomy until death, and the overall mortality is at least 50%[5]. There is still debate about whether there is any increased incidence of infection and death post-splenectomy[6].

The most common organism is *streptococcus pneumoniae* causing over 50% of infections[7]. Other organisms in decreasing frequency of infection include *haemophilus influenzae, neisseria meningitidis and e.coli.* No organism was isolated from case 3 although clinically and pathologically the diagnosis was not in doubt. Rarely in cases like this no organism is isolated, and the possibility of viral or other unidentified organism then arises.

Our case histories highlight several important points. All patients had a prodromal illness. If the patients and their families had understood the significance of urgent treatment of any prodromal symptoms, death might have been prevented. None were on prophylactic penicillin or had immediate access to antibiotics. They had not been vaccinated as splenectomy had been performed many years previously.

Opinion is divided over the use of prophylactic antibiotic therapy. Prophylactic penicillin does not always prevent overwhelming postsplenectomy infection as

sporadic case reports illustrate.[8,9] Zarrabi et al[10] reviewed all case-reports where patients were definitely stated to be receiving prophylactic penicillin. In only five out of fourteen did the patients have penicillin-sensitive pneumoccal infection. While the exact incidence of failure of penicillin prophylaxis cannot be calculated it would seem to be rare. Patient non-compliance must also be considered.

Traditionally penicillin has been used, but as up to 50% of infections are caused by non-pneumococcal organisms ampicillin has also been considered[3]. It would probably not be ethically acceptable to carry out a randomised long-term prospective study to decide the issue of prophylactic penicillin but retrospective data demands that the post splenectomy patient must be protected from overwhelming postsplenectomy infection as far as is possible.

The question of pneumococcal vaccination is also pertinent. Polyvalent pneumococcal vaccine is helpful in preventing pneumococcal infections in immunocompetent patients[11]. Post splenectomy the response to vaccination is impaired[12] but useful responses can be obtained[13]. Response to vaccination may also be impaired in the immunocompromised but some clinical effect is recognised.[14] Although the underlying disease and its treatment will affect the antibody response to vaccination these are not contra-indications. No vaccine is 100% effective. Pneumovax will not prevent infection with strains of pneumococci not represented therein.[12] Despite these reservations there are protective effects to be gained from vaccination. Current Department of Health guidelines recommend that pneumococcal vaccine is given, if possible, two weeks before splenectomy. Should vaccination prove impossible prior to surgery then it is recommended that it is carried out post splenectomy. Patients who have had a splenectomy and have never been vaccinated should be immunised at any opportunity that arises. In the splenectomised patient vaccination should be given every 5 to 10 years[14].

In cases of trauma, splenic salvage should be attempted where possible. Where operation is required, splenorrhaphy, partial splenectomy or heterotopic autotransplantation should be attempted[16].

Oral penicillin 250mg twice daily prophylactically for life is also advisable. In allergic patients erythromycin could replace penicillin. In these cases and those patients unlikely to comply with lifelong prophylaxis patient education is imperative. An informed discussion between patient, hospital doctor and general practitioner will help to ensure at least availability of an emergency antibiotic supply and awareness of the possible significance of prodromal symptoms. Wearing an engraved medic-alert bracelet may also be of value. In cases of trauma, conservation of any viable splenic tissue has a role. Increased awareness of the lifelong risk of splenectomy may help to avoid tragedies.

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